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(Citation)
Bulletin of health sciences Kobe,19:47-62

(Issue Date)
2004-03-25

(Resource Type)
departmental bulletin paper

(Version)
Version of Record

(JaLCDOI)
https://doi.org/10.24546/00391933

(URL)
https://hdl.handle.net/20.500.14094/00391933

KOBE UNIVERSITY

Secondary Functional Deterioration in Adults with Cerebral Palsy : Analysis of Developing Physiological Burnout

Toru Furui and Tomoaki Shimada

The purpose of this study is to investigate the development of secondary functional deterioration in middle aged adults with cerebral palsy (CP). The goal of the current study was to show that secondary disabilities are not inevitable for persons with CP. Subjects included 25 community dwelling adults with CP aged 41–69, and were classified into four groups of subjects by the change in the Barthel index (BI) score from 18 years of age (18 BI). Ambulatory (Type A) subjects did not deteriorate from a perfect BI score. Self–Cautious Type (Type S) subjects attained a perfect 18 BI score, and then slightly deteriorated. Non–ambulatory Bottoming out in Childhood Type (Type N) subjects experienced deterioration prior to the age of 18. Physiological Burn–out Type (Type PB) subjects deteriorated below 50% in the percentage ratio of the present score to the 18 BI score. In–depth investigation into the individual life history from childhood identified the key characteristics of subjects in each group responsible for their functional status. All subjects in the Type A group used a powered wheelchair from their adolescence. All Type PB subjects had exhausted their functional capacity by excess physical and mental exertion. It is suggested that over emphasis of the ability to independently walk from childhood possibly caused a delay in the use of wheelchairs in this group. It is concluded that to avoid possible physiological burnout, assistive technology and avoidance of excessive physical labor are recommended in persons with CP.

Key Words
Adult cerebral palsy,
Middle age,
Physiological burnout,
Secondary disability,
Barthel Index.

Introduction

Cerebral palsy (CP) is an unstable and sometime vulnerable condition with varying effects and influence throughout one's life. Secondary disabilities begin to threaten the ability of persons with CP to live independently. Accordingly, the goal of the present study was to firmly define the functional life cycle of individuals with CP. In addition, we wanted to identify some key factors associated with these individuals functional status. Specifically we wanted to determine why individuals with CP develop social integration while their health status fails to improve, and rather deteriorates. In addition, we wanted to determine the specific preventative measures aging persons with CP may take to attenuate the function loss in skill. Thus, we feel it important to clarify these issues in order to improve the rehabilitative approach to persons living with CP.

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Until recently, the primary clinical focus for persons with CP tended to be directed toward the care of young children[1]. However, within the last 15 years, issues concerning adults with CP began to attract greater societal focus[2-6]. This can also be considered an important human rights issue for people with disabilities. In 1995, Turk, et al. reported that adults with CP are facing serious clinical problems related to aging[7]. Pimm defined this issue of adults with CP as "physiological burnout"[8]. Physiological burnout may occur when persons with CP attempt to perform physical tasks beyond their capabilities over long periods of time. Eventually exhaustion of physiological systems occurs, resulting in a sharp decline in their functional abilities. Although this condition is not a new orthopedic discovery, but the number of these cohorts is increasing[9]. However, in the last 35 years, there has been no effective solution identified for preventing "physiological burnout" in adults with CP.

Japanese society's underdeveloped social systems for disabled persons affords a certain advantage with regard to conducting large-scale studies investigating secondary disabilities in community dwelling persons with CP. By the 1980 s, Japanese society had yet to equally accept persons with CP. In 1985 through surveys of 148 adults with CP living in Tokyo, Sato reported that "CP cannot be any longer considered a non-progressive disorder"[10]. Following these findings, in 1991, Ando and Ueda investigated 686 workers via a questionnaire who attended a workshop in Japan[11]. Most of these preceding studies were cross-sectional and based on questionnaire investigations[12-16]. Therefore, in the present investigation we utilized fieldwork skills that more closely reflect daily life to carefully define the life cycle of individuals with CP[17]. Our primary objective was to gather these data in order to reduce the threat of physiological burnout.

Subjects and Methods

1. Subjects and Procedures

Subject participants with CP older than 40 years of age were recruited from mainland Japan, were community dwelling, born prior to 1965 and ranged in age from 41 to 69 years of age. Twenty -five subjects (14 males, 11 females; mean age 51 years, SD 5 years 6 months) participated in this study (Table 1). Subjects consisted of four individuals who enrolled in a workshop for the physically handicapped, 17 were members of a few organizations for the handicapped of which one of the authors was connected to over the course of thirty years of field work. Two individuals were outpatients of a community rehabilitation unit where one of the authors had enrolled, and two individuals were members of an organization for secondary disabilities.

Subjects were fully informed of the goals and procedures involved in the study during the recruitment process and prior to each interview. Overall subjects were given telephone introductions. Our study prospectus and requests for cooperation were sent by mail only to those subjects who consented to participation via computer access on-line. After sending the letters, actual study consent was obtained in writing. Overall subjects fully cooperated in all aspects of this study. Interviews were carried out in a manner that was most convenient to the participants. Four subjects were interviewed at their workshop or at a public facility in

their community, and the other 21 interviews were conducted at the subjects' homes. This was convenient in gathering information on their childhood such as photos and videotapes. A semi-structured interview was performed face-to-face and audio tape-recorded. Because we wanted the participants to guide the interviews, the protocol was flexible and the order of discussion often shifted depending on the direction taken by each individual. When subjects lived with their parents or siblings, family members were also interviewed with participant's consent and presence. This study and documentation were provided from the viewpoint of a physical therapist. An outline of the components contained within the interview is as follows :

1) Therapeutic history following diagnosis with CP.
2) An accounting of the functional life history and each remarkable life event related to changes in motor function.
3) The best motor function status throughout life.

Researchers also performed a simple physical assessment, which evaluated deformity, muscle tone, pain, and other neuromuscular problems while performing daily living tasks. Seven participants were hospitalized for disabilities during their childhood and records were obtained through cooperation with the hospital. In all the participants, photos taken in their childhood were available and 12 subjects also provided us with motion picture data. Interviews were transcribed word for word and feedback was given to the interviewers. Individual chronological tables of the life history were made from all the gathered data and information.

2. Data Analysis

We compared individual mobility data by using the Gross Motor Functioning Classification System (GMFCS) with that of 5, 10, 15, 20, 30, 35, 40, 45, 50. 55, 60, and 65 year old subjects as well as that of subjects at the age of the present study[18]. Self-care data were translated into the Barthal index (BI) at the age of their best functional status, at 18 years of age, and at the time of study[19]. We felt it important to assess BI at 18 years of age due to the fact that contact with rehabilitation providers is reduced following high-school graduation, thus this is often a critical turning point in physical function.[5]

Changes of self-care in Activity of daily living (ADL) were estimated by the ratio of the present BI score(psBI)to the BI score at 18 years of age (18 BI). Furthermore the term "%18 BI" is used to refer to this ratio to calculate a percentage. We also compared the psBI and peak BI score (pkBI) by calculating the "%BI" percentage ratio of pkBI to psBI.

In addition, as a mobility item if a score of 5 points was obtained for "ability to operate more than 45 m with a wheelchair if unable to ambulate" originally in the BI, we added "ability to move more than 45 m by maneuver of a powered wheelchair" to it. We evaluated participants' present handicapped status by using the Craig handicap assessment and reporting technique (CHART), which was designed as a measure of long-term rehabilitation outcomes. CHART has five dimensions including physical independence, mobility, occupation, social integration, and economic self-sufficiency dimensions, with amaximum score of 100, and a maximum total CHART score of 500 points[20].

3. Subject type categorization

We wanted to consider these data not only to understand the individual life histories of subjects but also to group subjects into four types based on the BI score and its change (%18 BI). The maximum possible pkBI and psBI score is 100. Groups with a pkBI score of less than 95 were arranged according to the order of %18 BI. This categorization allowed us to see the pattern of physiological burnout in persons with CP. We classified the subjects as follows: Ambulatory Type (Type A) subjects had all the BI ratios and BI scores of 100. Self–Cautious Type (Type S) subjects had both pkBI and 18 BI scores of 100, in addition to a %18 BI of more than 50. Non–ambulatory Bottoming out in Childhood Type (Type N) subjects had a psBI, 18 BI and pkBI of less than 100, in addition to a %18 BI of more than 50. Physiological Burnout Type (Type PB) subjects had both a %BI and a %18 BI of less than 50.

Results

1. Diagnostic category

Athetosis was the most prevalent diagnostic category among subjects (n=16; 64%), and paralysis severity in these subjects was categorized as moderate in five subjects and severe in 11 subjects. Spastic diplegia was categorized as moderate in four (16%) subjects. Five subjects (20%) were diagnosed with spastic traplegia, one of these cases was severe and four cases were considered moderate. It is clear that athetosis was of the highest percentage in both the Type S (80%) and Type PB (71%) groups (see Table 1).

2. Health and functional status of subjects

In terms of deformities and contractures, Talipes equinus was documented in 12 cases, where limitations of range of motion or contractures of the knee were seen. Scoliosis was observed and documented in 11 cases (44%). However, four subjects exhibited severe "Windswept Hip Deformity"[21], the state in which the body axis is rotated with both legs bent out to one side, with one of these subjects having this condition from childhood. Atrophy within the deltoid or biceps brachii muscle was recognized in 14 cases over a moderate degree (56%). All 25 cases were accompanied by pain, except for one case of anesthesia on all extremities by progressive cord sign. This case had also undergone cystostomy. Neck, shoulder, and low back or pelvic pain were recognized in all pain complainants and remarkable coxalgia was observed in three subjects who were non–ambulatory and usually sat on the floor in a split sitting position (see Table 2).

Documentation of the outdoor ambulation method of subjects indicated drastic change over time. Only three subjects could walk independently at the time of the investigation. Thirteen subjects were able to walk independently in their best functional status, but 10 of these subjects lost this ability by the time of the investigation. In contrast, the total number of powered wheelchair users was 19 at the time of the investigation. Another six subject were pushed in a wheelchair by a caretaker whenever they would go out (see Table 3).

Eight of 25 subjects underwent cervical operations for secondary deterioration. They consisted in four Type S subjects and four Type PB subjects. There were three patients who received three separate operations each. The total number of operative interventions for secondary cervical myelopathy occurred 14 times in these eight subject (see Table 2).

3–1. Ambulatory Type (Type A)

Type A subjects include three partici-

Table 1. Characteristics of subjects

Groups		Total	Percentage	Ambulatory	Self-cautious	Non-ambulatory	Physiological Burnout
Age	Mean Age	51		48	54	51	50
	SD	5,52		6,65	5,43	6,75	1,53
Sex	Male	14	56	2	1	5	6
	Female	11	44	1	4	5	1
	Total	25		3	5	10	7
Athetosis	Moderate	12	48	2	4	2	4
	Severe	4	16			3	1
Spastic diplegia	Moderate	4	16	1	1	1	1
Spastic tetraplegia	Moderate	3	12			2	1
	Severe	2	8			2	
Education	No school	4	16			4	
	Specialjunior high school dropping out	1	4			1	
	Special juniorg high school	3	12			2	1
	Special senior high school	16	64	3	4	3	6
	Mainstream senior high school	1	4		1		
Living situation	live alone	6	24	1		2	3
	with partner	8	32	1	3	3	1
	with partner & children	6	24			4	2
	with partner & mother	1	4		1		
	with couple of brother	1 1	4		1		
	with a brother	1	4				1
	with a friend	1	4	1			
	with parents	1	4			1	
Marital status	Married (Mean±SD ; 30.6±4.18)	15	60	2	4	7	3
	Divorced	2	8			1	1
	Un-married	8	32	1	1	2	1
Children	3 children	1	4		1		
	2 children	4	16			2	2
	One child	6	24	1	1	4	
	No children	14	56	2	3	4	5
Type of job			duration	A	C	N	PB
	Office work job	1	6 year		1		
	Cleaning factory	4	3 year		2		2
	Ironworks	2	2 year				2
	Sawmill	1	2 year	1			
	Member of a City Council	1	7 year			1	
		9		1	3	1	4

Primary descriptive characteristics of the 25 subjects were various. Most subjects lived with a nuclear family and were married, 56% of subjects have no child, and 38% of subjects have work experience.

Table 2. Health and functinal status of each subject

Category	ID	Age	Age of Maximum function	Scores & Ratio of BI					Methods of going out at Maximum Function	Maximum Distance for Self Ambulation (km)	Powered Wheelchair use age	Degree of Involuntary Cervical Movement	Muscle atrophy of Deltoid/Biceps brachii	Low back pain	Pain of neck, shoulder and arm	Talipes Equinus	Knee Contracture	Scoliosis	Windswept Hip Deformity	Anesthesia	Coxalgia	Sitting on the floor in Split Sitting Position all day long.	Cervical surgery age	Operation frequency
				pk BI	% BI	18 BI	% 18 BI	ps BI																
A	A	50	26	100	100	100	100	100	IW	5	30	mod	1	4	3	0	0	2	0	0	0			
	B	41	23	100	100	100	100	100	IW	2	24	mod	0	2	3	0	0	0	0	0	0			
	C	54	22	100	100	100	100	100	IW	5	30	mod	2	3	3	0	0	0	0	0	0			
S	D	57	21	100	90	100	90	90	IW	7	46	mild	3	3	3	0	0	0	0	0	0		46	1
	E	62	36	100	90	100	90	90	IW	11	48	mild	3	3	3	0	0	0	0	0	0		39	3
	F	49	24	100	85	100	85	85	IW	5	48	mild	3	3	3	0	0	0	0	0	0		48	1
	G	52	19	100	80	100	80	80	AW	10	31	mild	1	2	3	0	0	0	0	0	0		46	3
	H	50	26	100	55	100	55	55	IW	1	42	mod	0	4	3	1	1	2	0	0	0			
N	I	46	14	65	38.5	20	125	25	AW	0.1	27	mild	3	3	3	4	4	4	4	0	4	4		
	J	50	19	80	100	80	100	80	Wc	3	32	mod	1	2	3	1	4	3	0	0	0			
	K	52	15	80	93.8	75	100	75	Wc	1	31	mod	1	2	3	4	4	0	0	0	0	2		
	L	47	16	70	57.1	40	100	40	Pc	0	32	mod	2	2	3	4	4	0	0	0	4	4		
	M	54	16	45	77.8	35	100	35	AW	0.5	33	mod	3	2	4	0	0	0	0	0	0	2		
	N	53	6	25	80	20	100	20	Pc	0		mod	1	3	4	4	4	4	4	0	0			
	O	69	57	65	92.3	65	92.3	60	Pc	0		seve	3	0	1	0	0	0	0	0	0			
	P	47	17	85	82.4	80	87.5	70	Aw	0.1	38	mild	2	4	4	1	4	0	0	0	0			
	Q	47	17	55	54.5	35	85.7	30	AW	0.7	35	mod	3	2	3	0	0	0	0	0	0	2		
	R	53	16	75	26.66	25	80	20	AW	0.8		mild	2	2	3	4	1	3	0	0	4	4		
PB	S	52	26	85	35.3	85	35.3	30	Wc	50	25	mod	4	2	4	4	4	0	0	0	0	2	52	1
	T	50	38	100	35	100	35	35	IW	12	39	mild	3	2	3	0	0	0	0	0	0		38	3
	U	50	26	70	28.6	70	28.6	20	IW	5		mild	4	1	3	0	0	3	0	0	0		42	1
	V	47	26	100	25	100	25	25	IW	25	43	mod	4	1	3	4	1	3	0	0	0			
	W	51	26	100	25	100	25	25	IW	20		mod	3	4	4	0	0	0	0	0	0	0	50	1
	X	51	21	100	15	100	15	15	IW	1.5	33	seve	4	4	4	4	1	3	4	0	0			
	Y	50	19	100	0	100	0	0	IW		20	mild	4	0	0	4	4	4	4	4	0			

In "Method of going out" items, abbreviations were as followings : IW ; Independent Walk, AW ; Assisted Walk by use of any assistive devices, such as a cane, crutch, walker, and human assistance. Wc ; manually self-propulsion of Wheelchair. Pc ; pushed in a wheelchir by caretaker.

Coding of categorical variables in grading of Deformity or Pain items were : 0 = no pain or deformity at all, 1=slightly, 2=mild, 3=moderate, 4=severe. Codeing of categorical variables in the "Split sitting" item were as follows : barely to change the position bu oneself=4, easy to move=2.

Table 3. Ambulation changes

		Electilic Powered Wheelchair present use				present Pushed in a wheelchair by caregiver	
		Type A	Type S	Type N	Type PB	Type N	Type PB
Ambulation Method at Maximum Function	Independent Walk	3	4		3		3
	Assisted Walk		1	4		1	
	self-propulsion of Wheelchair			2	1		
	Pushed in a wheelchair by caregiver			1		2	

Assisted walk refers to the use of any assistive device, such as a cane, crutch, walker, and human assistance.

pants, subjects A, B, and C (two males and one female) who had a full BI score of 100 throughout their life including the present. The average age, the diagnostic CP categories, marital status, and living situation of Type A subjects are shown in Table 1 and Table 2.

Within the Type A group, subject B is the youngest of all 25 participants at 41 years of age. Because of her young age, there is a high likelihood of experiencing further deterioration in the future. However, the lack of a deteriorated state of the subjects within the Type A group is very significant in understanding physiological burnout and its prevention. Three important similarities are found between these three subjects. First, all subjects were able to walk unassisted and were even able to run. Second, all subjects had begun living alone at a time when they were at their highest functional status. This event occurred at 26 years for subject A, 23 years for subject-B, and 22 years for subject C. Finally, all the subjects began using a powered wheelchair at 30 years of age for subject A, 24 years for subject B, and 30 years for subject C. The independent living of these subjects was supported by use of a powered wheelchair from the early stage until the present. It is important to note that this powered wheelchair usage was not due to an inability to ambulate, but used for convenience (for example when shopping) upon advice from other people cautioning them against overexertion.

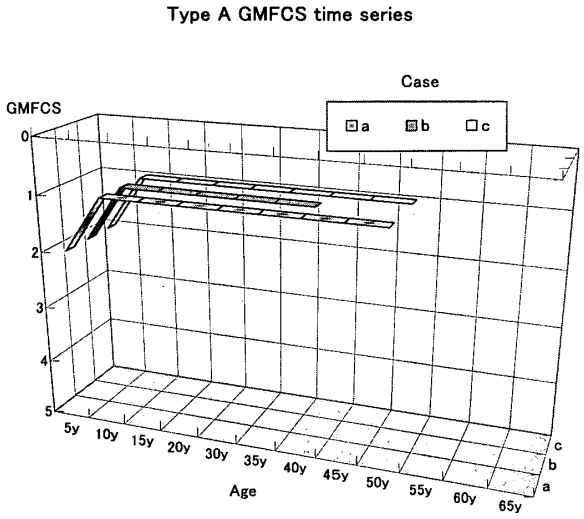


Figure 1. GMFCS level time series of Type A group subjects (subjects A, B, and C) show a maintenance of high levels (horizontal trend) after 10 years age.

GMFCS levels of the subjects within this group were very stable and high. Figure 1 illustrates the significance of the GMFCS levels of Type A group subjects, which demonstrated an almost horizontal graphical trend. The consistent GMFCS levels is a primary characteristic of the Type A group (see Figure 1).

3–2. Self-cautious Type (Type S)

The second group of subjects, self-cautious (Type S ; subjects D, E, F, G and H), attained an 18 BI of 100 and a psBI score around 80–90. However, the common characteristic among all members was that they were found to be careful and cautious regarding their physical condition and deterioration. All Type S group subjects except subject H were examined by orthopedists who were specialized in cervical operation caused by the sudden physical deterioration (see Table 2). Table 1 shows the diagnostic CP category, marital status, education, and work experience of Type S subjects.

From an analysis of the narrative life history, Type S subjects performed several intensive daily events as a result of physical overuse. Subject E worked for an organization that assisted wheelchair-bound individuals in climbing mountains. Subject G traveled extensively in Japan to give speeches and write articles as a representative of an organization for the handicapped despite being unable to barely stand with the assistance of a cane and required help to use a wheelchair. Subject F had been able to stand and run although her height was only 148 cm and weight was 48 kg, she was able to pull her 67 kg husband to his wheelchair and drive him around daily. Subject - D performed all household chores herself until her secondary disability occurred.

The Type S group consisted of one exceptional case and four subjects who received cervical surgical intervention. Two of the four subjects (subjects E and G) were involved with organizational groups for the handicapped. Due to their involvement, they had a greater knowledge and awareness of secondary disabilities associated with CP (cervical vertebral symptoms and the nature of myelopathy) at an early age than the other participants. Different physicians, using different techniques performed corrective surgical interventions for cervical dysfunction 3 times on these subjects at different times. These two subjects became more cautions with their bodies and made changes to their life-style following the surgical intervention. Subject E attempted to avoid overexertion and began to live with her brother and his wife in order to reduce the number of necessary household chores. She also began performing aquatic walking exercise. Subject G retired from the demanding front-line position within a handicapped organization and remained with the organization as a consultant and an advisor. This subject also commenced daily training and attended Qigong exercise class twice weekly.

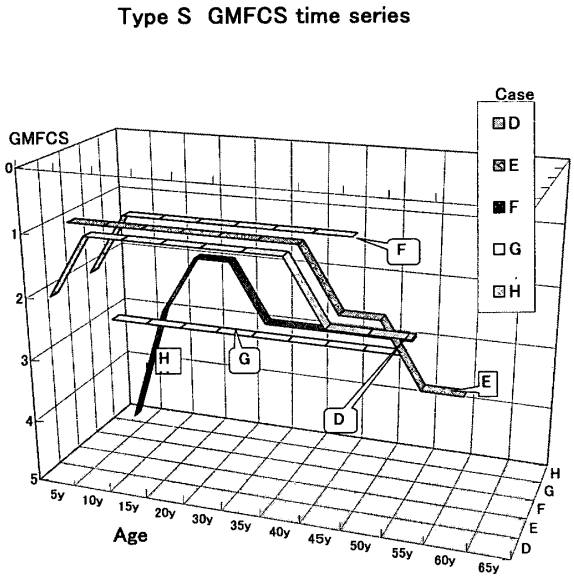


Figure 2. GMFCS level time series of Type S group subjects (subjects D, E, E, F, G, and H) show a concave downward trend with two dips when subjects were between 20 and 40 years of age.

Subject D also reduced the amount of housework she was doing by hiring more care-workers. Each time she felt tired, she rest for an hour. Subject F modified her physical exertion by living with a care worker to get assistance with the care of her husband who was much heavier than herself. This subject also began to receive thermotherapy and massage six times per week.

All of the five members of the Type S group realized they were at risk for further deterioration, and modified their life style accordingly. The change in GMFCS demonstrated a concave downward trend with two dips in subjects during their 20 s and 40 s (see Figure 2).

3-3. Non-ambulatory bottoming out in childhood Type (Type N)

The third subject group consists of Non-Ambulatory Bottoming Out in Childhood (Type N) and contains ten subjects (subjects I, J, K, L, M, N, O, P, Q, R). Type N subjects' pkBI was lower than 100. All ten Type N subjects had never been able to walk, and their physical change was minimal (the BI score decreased by approximately 10) after 18 years of age (see Table 2). The average age, gender ratio, diagnostic type, neurological impairment levels, academic history, and marital status are shown in Table 1. More subjects had been married in the Type-N group than in any of the other groups. Seven (70%) Type N subjects were married (as compared to 64% among all subjects), one was divorced, and only two participants had never been married. The academic history of the Type N subjects was shorter due to their heavier handicapped status. Three subjects finished high school, two attended special junior high school, one dropped out, and four had never attended school. Also, there was only one Type N subject (subject R) who had ever held a job. This subject has been working as a member of the municipal assembly for the past 4 years (See Table 1).

Thorough analysis of the life history reveals one of the most distinguishing characteristics of the Type N subjects; a critical event occurred at a young age to trigger a drastic physical change. At 6 years of age, subject-N experienced extensive hypertonia, including a rotation and side bending of his trunk caused by heavy ambulatory training. The hypertonia elicited major bodily dysfunctions at that time, after which the condition stabilized. Two subjects of the Type N group (subject I and subject P) participated in gait exercises with short leg braces on the parallel bar at around the age of 14. Subject I underwent brain surgery while subject P received knee surgery several times prior to experiencing serious bodily dysfunction. Following the deterioration of function, subject I's Activity of daily living (ADL) required complete assistance due to both upper and lower body hypertonia while subject P developed an inability to walk due to lower body hypertonia. Bodily dysfunction due to hypertonia was caused by both physical and mental changes associated with puberty in three subjects (subjects L, Q and R). Subjects Q and L used to be able to walk with a walker before the dysfunction, but had to use a wheelchair afterward. The ability to crawl following the dysfunction was found in subject L, in addition to the ADL level decreasing to a level requiring physical assistance. All of these changes occurred prior to 18 years of age. However, subject I's BI score increased after reaching 18 years of age, because of the onset of utiliza-

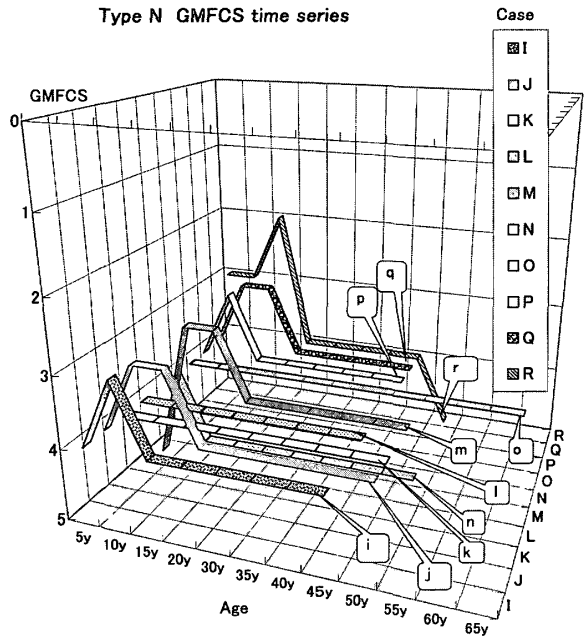


Figure 3. GMFCS level time series of Type N group subjects (subjects I, J, K, L, M, N, O, P, Q, and R) demonstrates a bottoming out followed by horizontal maintenance of this trend.

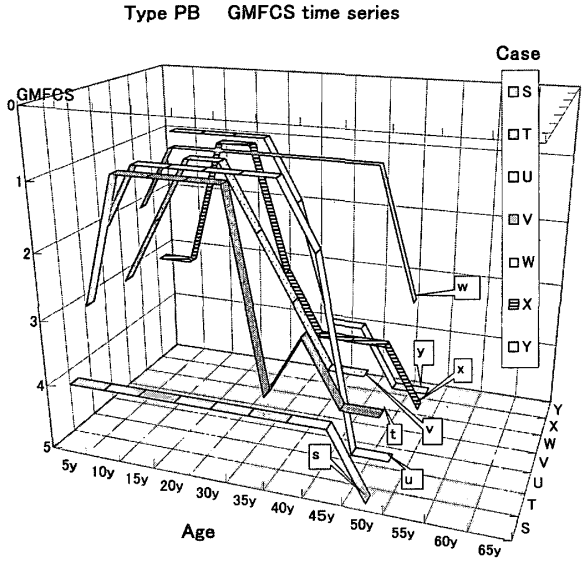


Figure 4. GMFCS level time series of Type PB ; Data demonstrate rapid drops in GMFCS levels in subjects S, U, V, W, X, and Y.
There are two sudden dips in subject T who underwent surgical care 3 times.

tion of a powered wheelchair rather than a change in bodily condition. Additionally, although subjects K, J, and O were experiencing difficulty standing because of knee contractures, they were of an infant status based on the %BI score, even though little dysfunction had occurred.

The change in the GMFCS level of most Type N subjects easily identifies the point of degradation occurring in the early years of life (see Figure 3).

3–4. Physiological burnout Type (Type PB)

The final group of subjects (n=7) were categorized as having physiological burnout (Type PB) and included subjects S, T, U, V, W, X, and Y. Physiological burnout (PB) occurs in individuals with CP when the locomotion system is weakened and damaged by the influence of CP and is then compounded by continuous exposure to additional stressors (Pimm 1992). According to Pimm, unlike psychological burnout, recovery is not possible in PB. However he also pointed out that the influence of PB could be mitigated or halted by appropriate case management and elimination of those factors, which lead to PB. Understanding the mechanisms involved in PB is a key to prevent and avoid secondary handicaps from occurring.

Most participants of the Type PB group were able to walk and had a pkBI score of 100 and a very low psBI score (35–0). The CP diagnostic category, gender, marital status, and education of Type PB subjects is shown in Table 2. Work history included two subjects who worked in a laundry store, two in ironworks, and three had never worked. The average working period was three years

SD one year and 8 months (see Table 2). Type PB subjects experienced rapid drops in GMFCS levels. (See Figure 4).

An examination of the historical data in Type PB subjects revealed several important trends. Subjects Y, V, T, and W of the Type PB group were the only four subjects who were employed. All four of the subjects' jobs were physically demanding. Two of these subjects (Y and V) were ironworkers who experienced serious accidents due to physical damage from overwork. After working a couple of years (subject Y, 3-years ; subject V 1-year) both participants realized that the bodily damage and dysfunction made it impossible to continue and they subsequently retired. However, both subjects continued to engage in activities that caused serious PB. Subject Y began involvement in and organization for the handicapped and became a caregiver to other disabled individuals. Subject V started helping perform housekeeping tasks in addition to engaging in extreme activities such as routinely walking home a distance of 25 km. Possibly these activities were responsible for eliciting PB.

Two subjects worked in a laundry shop for a period of time prior to retiring (subject W, 2-years ; subject T, 2-years). Subject W additionally bore the entire responsibility for caring for a dependent spouse and his two children. Due to this extreme work he is now experiencing secondary disabilities. Subject T began living with her mother-in-law following marriage with a physically dependant man. This situation was stressful and might have lead to secondary disabilities following the first cervical operation. Following this event subject T became divorced because she could not care for her dependent husband and work domestic jobs any more. After this divorce she began to participate in wheelchair soccer, which lead to a second and third operation.

4. Comparison in CHART scores

In the first study to evaluate CP by CHART, there was no significant relationship between pain and CHART scores[22]. Even if CHART scores had less validity in all aspects of pain, CHART is still considered a global measurement of disability. Therefore we used CHART scores to assess the outcomes of secondary disabilities in subjects with CP. Economic dimension scores of CHART did not show a statically significant difference between any of the groups. However total CHART scores were deferent

Table 4. CHART scores comparison between 4 groups

	Total CHART Scores			
	100-199	200-299	300-399	400-500
Physiological Burnout	3	2	2	
Non-ambulatory	1	2	6	1
Ambulatory			2	1
Self-cautious			3	2

Results of Spearman's correlation coefficient by rank test as follows :
Coefficient of correlation rs=0.678 (n=25), a meaningful point of coefficient of correlation ;
One side ($p<0.025$) both sides ($P<0.05$)=0.398, One side ($P<0.005$) both sides ($P<0.01$)= 0.511,
Z value=3.322, P value(both probability)=0.00089,
Same order rank correction Z value=3.104>2.576 [Z (0.995)],
Same order rank correction P value (both probabilty)= 0.0019<0.05,
Number of same order rank=8,

between the 4 groups (Type A, Type S, Type N, Type PB ; Table 4) There was a statistically significant difference ($P \leq 0.005$ in Spearman's correlation coefficient by rank test) among the 4 groups when we ranked total CHART scores into 4 groupings (below 200, 200–299, 300–399, 400–500). Type PB had the worst outcome of the 4 groups in spite of the fact that economic conditions were not poor. It is readily apparent from these results how strong an impact "physiological burnout" has on a person with CP.

Discussion

Occupation and accommodation support for persons with severe CP is minimal in Japan. The percentage of independent living in subjects in the current study was higher (92%) than Bottos's participants at only 12.5%, and Amdersson's at 87%. The percentage of participants who were married was significantly higher in our study (64%) than in any previous research. Bottos reported a marital ratio of 11.3%[12] and 35% of Andersson's participants were living with partner[13]. Judging from the above studies, despite the severity of neurological impairment participants in our study tended to be more active and outgoing than the majority of individuals with CP. The participants in our study had a rich life history full of variety and change. Because of this, our subjects might clearly demonstrate unique physical characteristics, tendencies, and problems.

Data in the current study were based on the level of change in GMFCS scores, change in BI, and an observation of the narrative life history of each subject. These data were analyzed to reveal three topics (deterioration, categorizing deteriorations, and prevention of deterioration) that are keys to management of life events in order to prevent physiological burnout. These three factors are discussed in the following section.

First, although Murphy reported that subjects demonstrated two peaks of physical deterioration in their 20 s and late 40 s, a third peak was identified that cannot be disregarded[2]. The newly revealed peak of deterioration in our study occurred around school age (5 to 10 years). The possible cause of deterioration at this point was the extreme physical preparation subjects underwent in order to attend school. Extreme physical training and intervention for complete incapability in standing or gait in young patients with severe CP could be responsible for causing excessive muscle tightness and systemic disorders. The chronic spasmodic hypertonus occurred when patients suffered severe pain by an accident, injury or an incorrect surgery. When there is no control over the cause of chronic pain, a painful experience in a person with CP is an easy invitation to physical degradation and depression of one's self–efficacy[23]. The developmental transition from toddler to childhood around school age, also represent a critical period for the prognosis of a person with CP.

Second, in the current study we classified subjects according to their %18 BI score. Despite the fact that BI is a scale of activity limitations according to the International Certification of Functioning Disability and Health (ICF)[24], diverse aspects of deterioration patterns were clearly observed in each category. Importantly, we were able to confirm that CHART scores reflected the pattern of results we obtained. Type N subjects were categorized as non–ambulatory at

18 years of age. Type N participants generally experienced a slow degradation in physical abilities following the first deterioration. On the contrary, even if subjects could walk at a high rate of speed (similar to a non-disabled pedestrian crossing the street), Type PB subjects had intensive deterioration when they continued doing daily activities beyond their ability. It is necessary, at this point, to explain that the cause of these secondary disabilities is not always related to neurological impairments. We feel this relationship may be clear as a result of our studies of subjects in the Type-PB group. The main cause of secondary disabilities in CP subjects (physiological burnout may be engagement in physical activities beyond their capabilities. Among these vigorous activities include walking at a rapid speed, care of other disabled parsons, hard manual labor over long periods of time (especially standing work). In our interview, subjects E, S, V, Y, W, F, T, and D unanimously agreed that they do not want to feel any less capable than individuals without disabilities. This competitiveness in CP subjects appears to have led them to overuse syndrome. Continuous stimulation of the powerless musculoskeletal system through simple activities such as standing and working in an individual with CP, can easily lead to physiological stress. As a typical example, the occipitofrontal diameter of the spinal canal in the neck of person with athetosis typed CP becomes narrowed[25-26]. It is assumed that involuntary movement and muscle spasms have enhanced this developing deformity in those persons with CP. In most cases, various motor disorders are easier to understand if you consider that CP is a progressive disorder rather than a stable condition.

Finally, although data from this study suggested that the secondary deterioration and physiological burnout could be prevented or delayed by understanding and using some effective preventative measures, the number of the subjects investigated in the present study is too small to apply statistical analysis. This point is a limitation of present study, and further quantitative investigations are required to identify detailed methodology for prevention of physiological burnout. Some hints for prevention of physiological burnout are illustrated in case of subject O, who suffered a very low functional deterioration, had excellent social outcomes, and was non-ambulatory. Because this subject understood the importance of relaxation and the avoidance of further physical stress, the degradation rate was reduced to only 93% at 69 years of age. This occurred despite sibling pressures throughout his life. Disabled individuals are urged to continue fitness regimes for not only health promotion but also prevention of secondary disabilities unless the fitness activities are too rigorous[27]. It is well known that the basal metabolism of an adult with CP is much higher than a normal individual[3], however little research has been done regarding these effects on the cardiovascular system and the relation to base physical fitness[28-29]. Thus, it is difficult to find appropriate standardized levels of required strength and fitness for individuals with CP. It was also reported that physical fatigue in adults with CP was not significantly associated with the type of CP and was most prevalent in persons with "self reported" moderate grades of CP[30]. This suggests that self-cognition of adults with CP reflects their over-use behavior and competitive attitudes. In fact, Tawada and coworkers reported that competitive

cases of athetiod CP tended to have progressive cervical changes as detected by MRI (Magnetic Resonance Imaging)[31].

Wheelchairs, in particular powered wheelchairs, are one of the most essential and efficient assistive technological products available to persons with CP. Many studies show that usage of a powered wheelchair during childhood helps individuals with CP develop better social and intellectual skills in addition to improving motivation and participation through providing a mechanism for mobility and independence[32, 33]. However, in all previous studies performed on adults with CP, those who could walk without support did not use a powered wheelchair[12, 13, 16]. The practical importance of using assistive technology products is emphasized through study of subject A, subject B and subject C. These subjects utilized a powered wheelchair from a healthy condition at a young age, even thought they could walk, and avoided physiological burnout. In the current study 13 of 25 subjects were previously able to walk with no assistant, but only three of these subjects could maintain this ability. In fact, 19 of 25 subjects used powered wheelchairs at the time of this investigation. Walking in children with CP is overemphasized and beyond realistic expectations, which often results in frustration by the individual when they are unable to attain this goal[12]. CP is too often thought of as a non-progressive disability, and in many instances preparations are not made by caregivers and parents of individuals with CP to accommodate the patient's changing function and needs[8]. Therefore, we strongly recommend that CP be regarded as a progressive disability.

To summarize, data analysis from the Type A group suggests that preventative maneuvers could be effective in delaying the onset of PB. The existence of cautious adults with CP, Type S, supports this hypothesis. The non-ambulatory group demonstrates a relationship between pediatric deterioration and a slower progression of dysfunction, similar to a functional bottoming out effect. However in the long term, these subjects have a better outcome than the physiological burnout (PB) group subjects in total CHART scores. Physiological burnout heavily impacted individuals with CP in self-care, mobility, and handicap. Maintenance of function is an essential issue of persons with CP, however there is little perception for the potential for deterioration to occur and little effective preparation for physiological burnout. We should rethink a traditional infant oriented rehabilitation approach to children with CP, which is primarily focused on the achievement of independent walking[12].

It is important that our society help the CP population view the use of assistive technological products, including powered wheelchairs as a preventative measure and not as a disgrace in seeking out physical assistance.

This paper is submitted as a doctoral thesis to Kobe University Graduate School of Medicine.

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